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A RARE CASE OF PRIMARY NASOETHMOIDAL MENINGIOMA

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ABSTRACT: Meningiomas of sinonasal tract is a rare entity especially in children. Primary sinonasal meningiomas often pose difficulty in diagnosis because of their infrequent occurrence and clinically they appear to be nasal polyp. The final diagnosis rests on the histological examination. Extra cranial meningiomas are rare in children and tend to be more aggressive. Here a case of primary ethmoid sinus meningoma with extension into the nasal cavity is presented and despite of its aggressive behavior it was benign. The importance of complete surgical extirpation is undoubted and results in a good survival rate. Histological grading of the tumour is not crucial in predicting the rate of recurrence. The paucity of reported cases is the evidence for its rarity.

Key Words: Meningioma, primary, sinonasal

INTRODUCTION

Meningiomas accounts for 10-15% of all intracranial tumours,^[1] but primary and secondary meningomas outside the central nervous system are uncommon. The most common localizations of extracranial meningiomas are the skull bones as well as scalp, nose, orbit, paranasal sinuses, middle ear, neck and skin.^[2]

Extra cranial meningiomas of the sinonasal tract are rare tumours and are frequently misclassified, resulting in inappropriate clinical management. Occurrence of primary extracranial meningiomas are probably related to the transformation of embryonic arachnoid cell remnants of ectopic meningocytes derived from pluripotent mesenchymal cells. To the best of our knowledge literature has 30 cases of primary meningiomas of paranasal sinuses. Meningiomas rarely occur in children, if so they may be present in association with Von Recklinghausen neurofibromatosis (type-I). Due to many clinical, topographical, radiological and surgical factors the histology is not solely decisive for the prognosis of the meningiomas.^[3] A genuine extracranial primary meningioma is to be confirmed after a CT scan, to rule out intracranial mass or any underlying bony erosion of the skull base. FNAC of the lesion can be deceptively mistaken and final diagnosis is usually made on the basis of histological examination of the excised mass. This extremely uncommon tumour justifies reporting an additional case, at the same time reviewing previous literature.

CASE REPORT

A thirteen year old male presented at the Department of ENT, Gandhi Medical College and Hamidia Hospital, Bhopal with complaints of swelling of the right side of face and proptosis of the right eye since 2 years, Nasal obstruction for 1 year and an episode of epistaxis 15 days back.

To start with patient's parents noticed prominence of the right eye which progressively increased over 2 years to the obvious forward protrusion of the right eye. Proptosis was painless and no difficulty in vision was reported by the patient (Figure 1). It was associated with swelling over the face on right side filling the nasofacial furrow. 1 year back patient developed blockage of right side of nose, which progressed to complete nasal obstruction. 15 days back patient had an episode of epistaxis from right side of nose, it was mild and bleeding stopped on its own.

Patient was of average built with lateral proptosis of the right eye and fullness of the right nasofacial groove. On anterior rhinoscopy pinkish polypoidal mass was seen completely filling the right nasal cavity pushing the septum to the left. Mass does not shrink on application of the decongestant, does not bleed on touch and on probing mass had attachment high up in the nasal cavity. Posterior Rhinoscopy revealed mass peeping through the choana.

Visual acuity and movements of the right eye were normal.

Neurological examination revealed no abnormality or cranial nerve palsy. The patient did not show clinical signs of neurofibromatosis.

Radiological examination of the paranasal sinuses revealed soft tissue shadow involving the right nasal cavity and the right maxilloethmoidal complex. CT Scan of the orbit and paranasal sinuses shows a soft tissue dense lesion with HO varying from +20 to +40 seen involving the right ethmoid sinus and right nasal cavity with gross expansion of the sinonasal cavity leading to lateral displacement of the medial wall of right orbit and anterolateral shift of the right eye ball (Figures 2 and 3). There was gross compression of right maxillary sinus with gross deviation of nasal septum to left and compression of the left



Figure 1: Patient with bulging right nasolabial furrow and proptosis of the right eye



Figure 2: CT scan coronal view showing homogeneously enhancing mass widening the sino nasal cavity and lateral bulging of the right eye

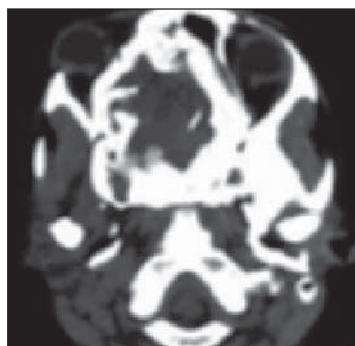


Figure 3: CT scan axial view



Figure 4: Photo micrograph showing spindle meningothelial cells and psammoma bodies (H/E, 100x)

nasal cavity. Lesion showed post contrast enhancement of the septa. There was no evidence of bony erosion and involved bones showed reactive sclerosis. There was complete opacification of the (right) frontal, maxillary and sphenoid sinuses suggestive of sinusitis. However optic canal and optic nerve on the involved side was normal. No intracranial connection or extension was noted.

Haemoglobin was 11.4g%, routine biochemistry and coagulation profile was normal. Patient was operated under General Anaesthesia on 20-1-05 by Lateral Rhinotomy approach. Moore's incision was given medial to the medial canthus of the (right) eye extending till (Right) Nasolabial fold. Mass was seen to be coming out from the ethmoid sinus and extending till lamina papyracea and cribriform plate. Mass was rubbery, friable in consistency and it was removed in piecemeal. The sinonasal cavity showed expansion but the walls were intact and there was no intracranial extension. The nasal septum was repositioned back to its place. The wound was closed in layers and nasal cavity was packed with medicated ribbon gauze. Post operative period was uneventful and mass of was sent for Histopathological examination.

The histopathological appearance was consistent with Psammomatous meningioma. Spindle cells with oval bland nuclei with many psammoma bodies and multifold calcification was seen (Figures 4). Mitosis was inconspicuous and no necrosis was seen.

Postoperative C.T. Scan showed no residual mass and patient is being followed on a monthly basis and shows no evidence of recurrence at 3 months postoperatively.

DISCUSSION

Extra cranial primary meningioma is notably a tumour of rare occurrence with an incidence of 0.9% as compared to 2% seen in the case of secondary meningiomas.^[4] Such a tumour occurring in the paranasal sinus is believed to arise from arachnoid 'cap cells' or meningocytes, which have migrated in the nerve sheath and become detached during development.^[5] The classification system of Hoye^[6] encompasses the major etiologies proposed in the development of extracranial meningiomas.

1. Extracranial extensions of a meningioma with an intracranial origin (Secondary).
2. Extracranial extensions of a meningioma arising in a neural foramina (Primary).
3. Ectopic, without any connection either to foramen of a cranial nerve or to intracranial structures (Primary).
4. Extracranial metastasis from an intracranial meningioma (Secondary).

Our case belonged to the third group as there was no clinical or radiological evidence of an intracranial lesion.

The current WHO classification distinguishes 3 grades of meningioma the typical or benign type (Grade I), the atypical with frequent mitosis (Grade II) and the anaplastic type with invasion (Grade III). However Zulch^[7] stated that it is not the histological grading which is most crucial in the rate of recurrence of meningiomas, but primarily the completeness of extirpation.

Meningiomas are benign in so far as they do not metastasize, but they often show a predilection for local permeation of crevices and foramina, whilst pressure necrosis may result in spread from one cavity to other.^[8] In our case benign nature of the tumour was evident from the slow growth of the tumour and non involvement of the orbit or any cranial nerves.

Meningiomas are rare in children but however these tend to be more aggressive in growth, size, propensity to undergo malignant changes and recurrence rate among children. CT Scan or MRI precisely gives information of the extent and invasion of the tumor and is imperative in the diagnosis. However, the final diagnosis depends on the histological examination. Hyperostosis of surrounding bone a classic finding in meningioma was also noted in our case.

76% of tumours have progesterone receptors, 96% have somatostatin receptors, 89% have Epidermal growth factor receptors, 19% have estrogen receptors. Hence the use of Tamoxifen and RU-486 (antiprogesterone) are under study. (Bradley 1273). Characteristic and most frequent chromosomal aberration in meningiomas is monosomy 22, which however, has been shown not to be relevant to the prognosis.^[9]

Radical surgical resection remains the primary mode of treatment and is correlated with a good prognosis. Efficacy of adjunctive radiotherapy after surgery is not established.^[1] Radiotherapy is used in unresectable malignant meningioma or recurrent meningioma where surgery is not feasible. Newer techniques in treatment include proton irradiation and stereotactic radiosurgery with gamma knife.

Chances of recurrence of meningioma are increased in

cases of –

- Incomplete surgical removal.
- Atypical and malignant histologic types.
- Presence of nucleolar prominence.
- Presence of more than 2 mitosis per 10 high power field.
- Heterogenous tumour contrast enhancement on CT Scan.

In general, prognosis following surgical resection is good (72% at 5 years).

The diagnosis and management of this tumours is particularly important because of their infrequent occurrence and unpredictable clinical behavior. A clear understanding of etiology and appropriate diagnostic and management principles helps to overcome the challenges posed by primary extracranial meningiomas.

SUMMARY

- A case of primary Naso ethmoidal meningioma is presented that is extremely rare in children.
- It is often confused with Nasal polyp and final diagnosis rests of the histological examination of the excised mass.
- CT scan is imperative in diagnosis, extent of the disease and to rule out intracranial extent of the mass.
- Complete surgical removal is the definite treatment and so also the good prognostic indicator.

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